Mass General Brigham
Health Plan

Uptravi® (selexipag)
Effective 03/01/2023

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☒ Medical Benefit		
Specialty Limitations	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
Contact Information	**Medical and Specialty Medications** All Plans | Phone: 877-519-1908 | Fax: 855-540-3693		
	Non-Specialty Medications All Plans | Phone: 800-711-4555 | Fax: 844-403-1029		
Exceptions	N/A		

Overview
Uptravi® (selexipag) is indicated for the treatment of pulmonary arterial hypertension (PAH, WHO Group I) to delay disease progression and reduce the risk of hospitalization for PAH.

Coverage Guidelines
Authorization may be reviewed for members new to the plan who are currently receiving the requested medication excluding when the product is obtained as samples or via manufacturer's patient assistance program.
OR
Authorization may be granted for members when all the following criteria are met, and documentation is provided:

1. Member has PAH defined as WHO Group 1 class of pulmonary hypertension
2. PAH was confirmed by **ONE** of the following:
    a. Pretreatment right heart catheterization with **ALL** of the following results:
        i. mPAP >20 mmHg
        ii. PCWP ≤15 mmHg
        iii. PVR ≥3 Wood units
    b. For infants less than one year of age, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.

<u>Continuation of Therapy</u>
Reauthorization requires physician documentation of continuation of therapy for members who are experiencing benefit from therapy as evidenced by disease stability or disease improvement.

Limitations
1. Initial approvals and reauthorizations will be granted for 12 months.
2. The following quantity limits apply:

Uptravi 1000 mcg, 1200 mcg, 1400 mcg, 1600 mcg, 400 mcg, 600 mcg, 800 mcg	60 tablets per 30 days
Uptravi 200 mcg	140 tablets per 28 days
Uptravi Therapy Pack	1 pack per 28 days

Appendix

WHO Classification of Pulmonary Hypertension

1 PAH

1.1 Idiopathic (PAH)
1.2 Heritable PAH 1.3 Drug- and toxin-induced PAH
1.4. PAH associated with:
- 1.4.1 Connective tissue diseases
- 1.4.2 HIV infection
- 1.4.3 Portal hypertension
- 1.4.4 Congenital heart diseases
- 1.4.5 Schistosomiasis

1.5 PAH long-term responders to calcium channel blockers
1.6 PAH with overt features of venous/capillaries (PVOD/PCH) involvement
1.7 Persistent PH of the newborn syndrome

2 PH due to left heart disease

2.1 PH due to heart failure with preserved LVEF
2.2 PH due to heart failure with reduced LVEF
2.3 Valvular heart disease
2.4 Congenital/acquired cardiovascular conditions leading to post-capillary PH

3 PH due to lung diseases and/or hypoxia

3.1 Obstructive lung disease
3.2 Restrictive lung disease
3.3 Other lung disease with mixed restrictive/obstructive pattern
3.4 Hypoxia without lung disease
3.5 Developmental lung disorders

4 PH due to pulmonary artery obstruction

4.1 Chronic thromboembolic PH
4.2 Other pulmonary artery obstructions
- 4.2.1 Sarcoma (high or intermediate grade) or angiosarcoma
- 4.2.2 Other malignant tumors
  - Renal carcinoma
  - Uterine carcinoma
  - Germ cell tumours of the testis
  - Other tumours
- 4.2.3 Non-malignant tumours
  - Uterine leiomyoma
- 4.2.4 Arteritis without connective tissue disease

4.2.5 Congenital pulmonary artery stenosis
4.2.6 Parasites
Hydatidosis

5 PH with unclear and/or multifactorial mechanisms
5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders
5.2 Systemic and metabolic disorders: Pulmonary Langerhans cell histiocytosis, Gaucher disease, glycogen storage disease, neurofibromatosis, sarcoidosis
5.3 Others: chronic renal failure with or without hemodialysis, fibrosing mediastinitis
5.4 Complex congenital heart disease

References
1. Uptravi [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; September 2019.
2. Sitbon O, Channick R, Chin K, et al. Selexipag for the treatment of pulmonary arterial hypertension. N Engl J Med. 2015;373:2522-33.
3. Simonneau G, Robbins IM, Beghetti M, et al. Updated clinical classification of pulmonary hypertension. J Am Coll Cardiol. 2013;62:D34-S41.
4. Rubin LJ; American College of Chest Physicians. Diagnosis and management of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Chest. 2004;126(1 Suppl):7S-10S.
5. McLaughlin V, et al. ACCF/AHA 2009 Expert Consensus Document on Pulmonary Hypertension. J Am Coll Cardiol. 2009;53:1573-1619.
6. Klinger, JR., Elliott, CG, Levine, DJ, et al. Therapy for Pulmonary Arterial Hypertension in Adults: Update of the CHEST Guidelines and Expert Panel Report. Chest. 2019:155(3): 565-586.
7. Galie, N., McLaughlin, VV, Rubin, LJ, Simonneau, G. An overview of the 6th World Symposium on Pulmonary Hypertension. Eur Respir J 2019; 53: 1802148; DOI: 10.1183/13993003.02148-2018. Published 24 January 2019.
8. Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. Eur Respir J 2019;53:1801913; doi:10.1183/13993003.01913-2018.

Review History
11/16/22 – Switched to custom criteria. Matched SGM. Effective 03/01/2023.